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A Case of Arteriovenous Type Cardiac Hemangioma

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Cardiac hemangiomas are rare primary tumors of the heart and constitute only 2.8% of primary cardiac tumors. They are classified into capillary, cavernous, epitheloid and arteriovenous type and the last one is the most uncommon type. We experienced a case of cardiac hemangioma which was diagnosed as arteriovenous type for the first time in Korea in the literature. The patient was a 54-year-old woman who presented with palpitation and anterior chest pain. The diagnosis was based upon coronary angiography which showed two tumor blushings located in the interatrial and interventricular septum with venous drainage to the coronary sinus and right atrium. Associated atrial fibrillation with rapid ventricular response was controlled with digitalis.

Key Words : *Cardiac hemangioma, Arteriovenous type, Atrial fibrillation*

INTRODUCTION

Hemangioma is one of the most common soft tissue tumors comprising 7% of all benign tumors[1]. The majority of hemangiomas are superficial lesions that have a predilection for the head and neck region, but they may also occur internally, notably in organs such as the liver. But cardiac hemangiomas have been known as very rare tumors located in any part of the heart, including endocardium, myocardium and epicardium[2].

Hemangiomas are classified histologically into capillary, cavernous, arteriovenous and epitheloid type and, among them, capillary and cavernous types are known to be encountered more frequently. According to the literature[3], the same classification can be applied to cardiac hemangioma.

Arteriovenous type which is characterized pathologically by arteriovenous malformation and radiologically by venous drainage during the arterial phase is a very rare type of cardiac hemangioma. We experienced a case with arteriovenous type of cardiac hemangioma which was diagnosed by coronary angiography for the first time in Korea. We report our case with a review of the literature on cardiac hemangiomas.

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CASE REPORT

A 54-year-old woman was admitted to our hospital complaining of intermittent palpitation which began 6 years ago and aggravated during the last 2 months. After the first symptom, she visited a local clinic and her symptom was managed under the impression of cardiac arrhythmia. But one year later, she discontinued anti-arrhythmic medication on her own. Until two months before admission, she had experienced intermittent palpitation again but it did not interfere with her life. Afterwards, palpitation became more severe suddenly and squeezing anterior chest tightness radiating to the left axilla developed and continued for about 10 minutes.

The patient was a housewife and had no history of smoking or alcohol consumption. There was no specific problem in her past medical and family history. The body temperature was 36.5℃ and the heart beats were

irregularr and rated over 140/min. On physical examination, mild moist crackling sounds were heard below a third on the back bilaterally. Neck vein engorgement was also seen. Considering the chest pain as ischemic in nature, nitroglycerin was given sublingually and the pain subsided. ECG showed atrial fibrillation with rapid ventricular response. Cardiomegaly with pulmonary vascular blurring and redistribution was seen on chest X-rays (Fig. 1). Other laboratory data, including cardiac enzymes and thyroid function tests, were within normal limits. Initially, cardiac tumor was not considered as the differential diagnosis. Transthoracic echocardiography, which was performed after her symptom had disappeared, showed normal global left ventricular systolic function (ejection fraction : 63%) with left ventricular enlargement and hypertrophy, and also showed trivial mitral regurgitation. On coronary angiogram, the left coronary arteries showed normal findings. But in the right coronary artery, we could find two tumor blushings which were supplied by a conus branch (Fig. 3). The one was on the right atrial side of the interatrial septum draining directly to the right atrium and the other was on the interventricular septum communicating with the coronary sinus via a large vein. The venous drainages of the tumor blushings could be seen simultaneously during the arterial phase. These two tumor blushings were measured about 18x10mm and 44x9.8 mm in length, respectively. These angiographic findings were compatible with the arteriovenous type of cardiac hemangioma. Left ventriculography and MRI failed to define the tumor masses. Retrospective video review of transthoracic echocardiography revealed no echogenic mass around the interatrial septum. We concluded that the patient had cardiac hemangiomas associated with atrial fibrillation

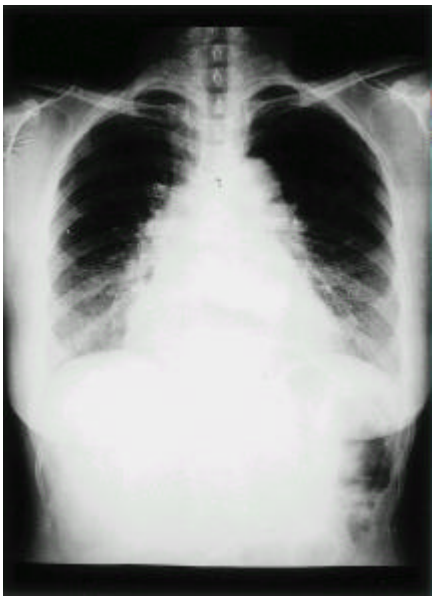

Fig. 1 Chest x-ray showed cardiomegaly with pulmonary vascular blurring and redistribution.

DISCUSSION

Primary tumors of the heart are uncommon and, among them, cardiac hemangioma is an exceptionally rare one. According to the report of MaAllister et al.[2], the incidence of cardiac hemangioma in autopsy was varying

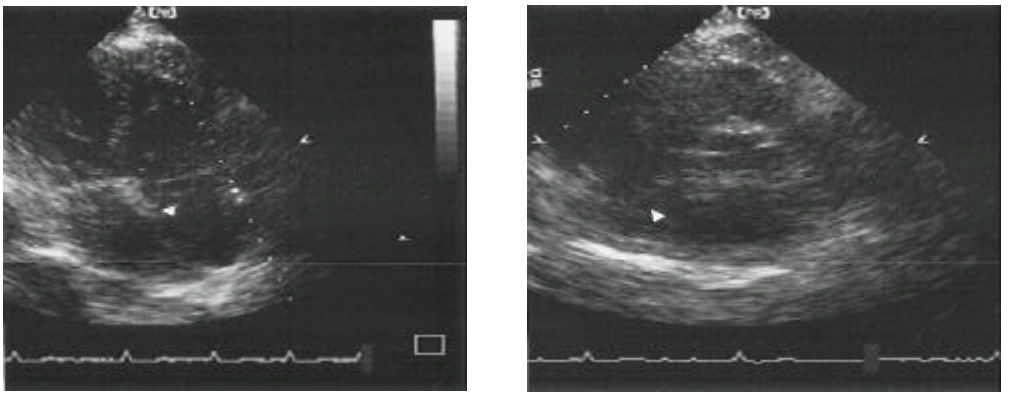

Fig. 2 Retrospective review of transthoracic echocardiography with relatively poor acoustic windows showed no echogenic mass around interatrial septum (arrowhead). **Right :** parasternal short axis view. **Left :** apical four chamber view.

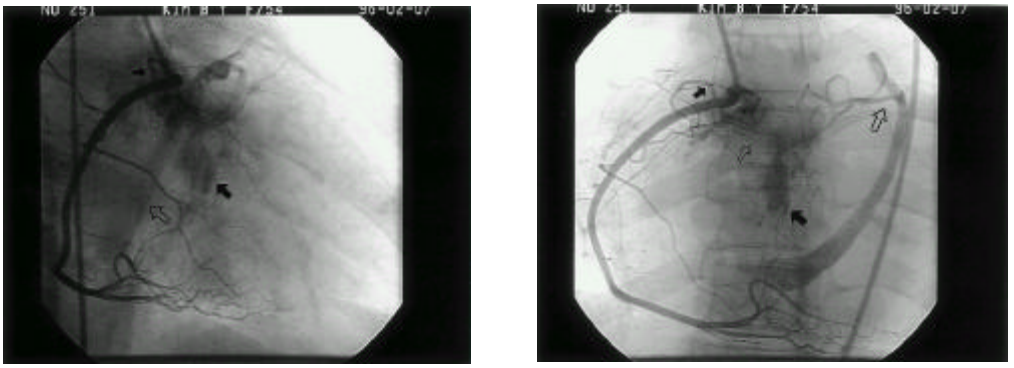

Fig. 3. In the right coronary angiography, there were two tumor blushings which were supplied by a conus branch.(small arrow) **Left :** one was on the right atrial side of the interatrial septum (small open arrow) and drained directly to right atrium. **Right :** the other was on the interventricular septum(large arrow) communicating with coronary sinus via large veins(large open arrow). Both of the venous drainages could be seen simultaneously in the arterial phase. These findings were compatible with arteriovenous type of cardiac hemangiomas.

between 0.0017% and 0.27%. Others also reported that the incidence at autopsy ranges from 0.001% to 0.03%[4]. Among the benign cardiac tumors, myxoma is the most common tumor constituting more than 50% of cases. Other benign cardiac tumors are rare and include fibroma, lipoma, rhabdomyoma, hemangioma, papillary fibroelastoma, teratoma, mesothelioma of the atrioventricular node, granular cell tumor, neurofibroma and lymphangioma. Cardiac hemangioma constitutes only 2.8% of primary cardiac tumors[2].

The first case of cardiac hemangioma which was located in the left atrium was reported by Uskoff in 1893[5]. But the exact number of previously reported cases was uncertain because cardiac hemangioma had been included under the titles of vascular hamartoma, angioma or cardiac varices in the past. Prichard reported as many as 46 cases in 1951[6], yet recently Brizard et al reported 23 cases in 1993[3].

Cardiac hemangioma can occur at any age and symptoms depend on the anatomic situations. According to Brizard et al[3]., among the 23 cases the most frequent clinical presentation was dyspnea on exertion (43%, 10/23). Less frequently, patients presented with arrhythmias (17%, 4/23), pseudoangina (13%, 3/23), signs of right heart failure (13%, 3/23), pericarditis or pericardial effusion(8%, 2/23) and failure to thrive (8%, 2/23).

Cardiac hemangiomas can occur in the endocardium, myocardium or epicardium. According to Brizard et al., they are commonly located on the lateral wall of the left ventricle and on the anterior wall of the right ventricle[3]. Intramural growths may be responsible for atrioventricular block[6] and compression of the coronary vessels can mimic myocardial infarction. A subendocardial hemangioma located on the right side of the interventricular septum may simulate right ventricular outflow tract obstruction[7-11]. Tumors developing in the pericardium can result in pericardial effusion, cardiac tamponade and renal failure[12]. When the cardiac hemangioma is growing in the left atrium, a picture of mitral regurgitation, due to mitral valve prolapse associated with fatigue and dyspnea, can occur[13].

Just like hemangioma from the other sites of the body, cardiac hemangioma are also composed of a benign proliferation of endothelial cell and are classified into capillary, cavernous, epitheloid and arteriovenous types[1]. Among them, arteriovenous hemangioma can be divided into two subtypes: one is that which occurs in various internal organs and which is associated with varying degrees of arteriovenous shunt; the other is that which occurs superficially in the dermis which is not associated with significant arteriovenous shunt. The former is a much more important type and it is regarded by some authors as an arteriovenous malformation. It can be conceptualized as partial persistence of the fetal capillary bed, resulting in anomalous connection between the arteries and the veins. Distinctive gross appearance of this lesion has earned various names, including *cirsoid aneurysm, racemose hemangioma and arteriovenous aneurysm*.

Arteriography is indispensable in the diagnosis of these tumors because it demonstrates large tortuous vessels of both the arterial and venous types with early filling of the draining veins during the arterial phase.

In the diagnosis of cardiac hemangioma, echocardiography, cardiac catheterization, coronary angiography, computed tomography and MRI are available. Two-dimensional echocardiography will show an echogenic mass lesion that contains echolucent areas[14]. These lucencies represent the dilated vascular channels or cavernous lakes found in hemangioma. With the advance of transesophageal echocardiography, assessment of cardiac mass, especially in the cardiac base, has significantly improved[15]. Furthermore, color flow Doppler imaging, especially with emphasis on the lower velocities, can delineate clearly the flow around the echolucent mass and substantiate its presence further[14]. Computed tomography in cardiac hemangioma most likely will be interpreted as normal because of the limitation due to the poor resolution created by the motion of the heart. Using contrast medium with MRI has been reported to delineate the vascularity of these tumors by showing intense contrast enhancement with tumor calcification and flow voids[16]. According to a recent case report, definitive diagnosis of cardiac hemangioma could be made preoperatively by gadolinium enhanced MRI alone[17]. Cardiac catheterization can localize a tumor within the atrium or the ventricle and coronary angiography can outline a patch of neovascularity, so called tumor blush. It is conversely worth mentioning that long-standing left atrial thrombosis in rheumatic mitral disease may also be revealed as a tumor blush especially from the left coronary injection. Likewise, myxoma can be supplied from the coronary arteries which can develop tumor blush and mimic hemangioma[13].

Hemangiomas of the heart, even when recognized preoperatively, are not always amenable to surgery. Successful resection can be performed only when the hemangioma is well circumscribed and small. Surgical resectability is evaluated with computed tomography, MRI or coronary angiography[3].

The natural history of cardiac hemangioma is unpredictable. They may involute[18], cease growing or proliferate indefinitely but remain benign and silent until the patient dies of an untreated cause. Some investigators reported that, even when complete resection is not performed, long term outcome in these patients seems to be favorable[3]. Others reported that the prognosis for unresectable hemangioma must be poor[13]. In inoperable symptomatic cases, irradiation and cardiac transplantation can be considered though this has not been reported.

REFERENCES

1. Eizinger FM. *SOFT TISSUE TUMORS. 3rd Edition. St. Louis : Mosby-Year Books, Inc, 1993.*
2. MaAllister HA, Fenoglio JJ. *Tumors of the cardiovascular system. In Atlas of Tumor,Patholgy.Fascle 15, second edition. Washington D.C : Armed Forces Institute of Pathology, 1978.*
3. Brizard C, Latremouille C. *Cardiac Hemangioma. Ann Thorac Surg 1993;56:390-394.*
4. Straus R, Murliss R. *Primary tumors of the heart. Arch Pathol Lab Med 1945;39:74-78.*
5. Sulayman R, Cassels DE. *Myocardial Coronary Hemangiomatous Tumors in Children. Chest 1975;68:113.*
6. Prichard RW. *Tumors of the heart. Arch Pathol 1951;51: 98-128.*
7. Weir I, Aills P. *A case of left atrial hemangioma: echocardiologic, surgical and morphological feature. Br Heart J 1987;58:565-668.*
8. Cho SO, Kim SL. *A case of Cardiac Hemangioma Causing RVOT Obstruction. J Int Med 1991;38;262-267.*
9. Soberman MS, Plauth WH. *Hemangioma of the right ventricule causing outflow tract obstruction. J Thorac Cardiovasc Surg 1988;96;307.*
10. Nicks R. *Hamartoma of the right ventricle. J Thorac Cardiovasc Surg 1964;47:762.*
11. Delaye J, Crassard P. *Les tumeurs primitives du ventricule droit, A propos dun cas dhémangio-endothéliome bénin opré avec succés. L Med Lyon 1972;47:483.*
12. Stoupel E, Primo G. *Cardiac tamponade with renal failure due to hemangioma of the heart. Acta Cardiologica 1979; 5:345-351*
13. Chao JC, Reyes CV. *Cardiac Hemangioma. Southern Medical Jounal 1990;83:44-47*
14. Cunningham T, Lawrie GM. *Cavernous Hemangioma of the Right Ventricle: Echocardiographic-Pathologic Correlates. J Am Soc Echocardiogr 1993;6:335-340*
15. Mugge A, Daniel WF. *Diagnosis of non-infective cardiac mass lesions by two-dimensional echocardiography. Circulation 1991;83:70-81*
16. Jennifer L, Randolph M. *MR and CT Appearance of Cardiac Hemangioma. Journal of Computer Assisted Tomography 1996; 20(3);482-483*
17. Fukunzawa S, Yamamoto T. *Hemangiomas of the left ventricular cavity. Heart Vessels 1993;82 11-214.*
18. Palmer TE, Tresch DD. *Spontaneous resolution of a large cavernous hemangioma of the heart. Am J Cardiol 1986; 58:184-185*